

wherein: X^+ is $N^+(R_1, R_2, R_3)$, wherein

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R_1, R_2, R_3 , being the same or different, are selected in the group consisting of hydrogen, a C_1 - C_9 straight or branched alkyl group, $-CH=NH(NH_2)$, $-NH_2$, and $-OH$; or one or more R_1, R_2 and R_3 , together with the nitrogen atom which they are linked to, form a saturated or unsaturated, monocyclic or bicyclic heterocyclic system; with the proviso that at least one of the R_1, R_2 and R_3 is different from hydrogen;

Sub C1

Z is selected from

Sub C1

- $-OR_4$,
- $-OCOOR_4$,
- $-OCONHR_4$,
- $-OCSNHR_4$,
- $-OCSOR_4$,
- $-NHR_4$,
- $-NHCOR_4$,
- $-NHCSR_4$,
- $-NHCOOR_4$,
- $-NHCSOR_4$,
- $-NHCONHR_4$,
- $-NHCSNHR_4$,
- $-NHSOR_4$,
- $-NHSONHR_4$,
- $-NHSO_2R_4$,

-NHSO₂NHR₄, and

-SR₄,

wherein -R₄ is a C₁-C₂₀ saturated or unsaturated, straight or branched alkyl group, optionally substituted with an A₁ group, wherein A₁ is selected from the group consisting of a halogen atom, or an aryl, heteroaryl, aryloxy or heteroaryloxy group, said aryl, heteroaryl, aryloxy or heteroaryloxy groups being optionally substituted with one or more C₁-C₂₀ saturated or unsaturated, straight or branched alkyl or alkoxy group and/or halogen atom;

Y⁻ is selected from the group consisting of -COO⁻, PO₃H⁻, -OPO₃H⁻, tetrazolate-5-yl;

with the proviso that when Z is -NHCOR₄, Y is -COO⁻, then R₄ is C₂₀ alkyl;

with the proviso that when Z is -NHSO₂R₄, Y⁻ is -COO⁻, then R₄ is not tolyl;

with the proviso that when Z is -NHR₄, X⁺ is trimethylammonium and Y⁻ is -COO⁻, then R₄ is not C₁-C₆ alkyl,

their (R,S) racemic mixtures, their single R or S enantiomers, or their pharmaceutically acceptable salts .

2 29. (New) A compounds according to claim 28, wherein R₁, R₂ and R₃ are methyl.

30. (New) A compounds according to claim 28, wherein the heterocyclic system formed by R₁, R₂ and R₃ together with nitrogen is selected from the group consisting of morpholinium, quinuclidinium, pyridinium, quinolinium and pyrrolidinium.

31. (New) A compound according to claim 28, wherein R₁ and R₂ are H, R₃ is selected from the group consisting of -CH=NH(NH₂), -NH₂ and -OH.

32. (New) A compound according to claim 28, wherein Z is selected from the group consisting of ureido (-NHCONHR₄) or carbamate (-OCONHR₄), and R₄ is a C₇-C₂₀ saturated or unsaturated, straight or branched alkyl group.

33. (New) A compound according to claim 32, wherein R₄ is a C₉-C₁₈ saturated or unsaturated, straight or branched alkyl group.

34. (New) A compound selected from the group consisting of

R,S-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;

R,S-4-quinuclidinium-3-(tetradecyloxycarbonyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(nonylcarbamoyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(nonyloxycarbonyl)-oxybutyric acid chloride;

R,S-4-trimethylphosphonium-3-(nonylcarbamoyl)-oxybutyrate;

R,S-4-trimethylammonium-3-(octyloxycarbonyl)-aminobutyrate;

R,S-4-trimethylammonium-3-(nonyloxycarbonyl)-aminobutyrate;

R,S-4-trimethylammonium-3-octyloxybutyrate;
R,S-4-trimethylammonium-3-tetradecyloxybutyrate;
R,S-1-guanidinium-2-tetradecyloxy-3-(tetrazolate-5-yl)-propane;
R,S-4-trimethylammonium-2-tetradecyloxy-3-(tetrazolate-5-yl)-propane;
R,S-3-quinuclidium-2-(tetradecyloxycarbonyl)-oxy-1-propanephosphonate
monobasic;
R,S-3-trimethylammonium-2-(nonylaminocarbonyl)-oxy-1-
propanephosphonate monobasic ;
R,S-3-pyridinium-2-(nonylaminocarbonyl)-oxy-1-propanephosphonic acid
chloride;
R-4-trimethylammonium-3-(tetradecylcarbamoyl)-aminobutyrate;
R-4-trimethylammonium-3-(undecylcarbamoyl)-aminobutyrate;
R-4-trimethylammonium-3-(heptylcarbamoyl)-aminobutyrate;
R,S-4-trimethylammonium-3-(nonylthiocarbamoyl)-aminobutyrate;
R-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;
S-4-trimethylammonium-3-(nonylcarbamoyl)-aminobutyrate;
S-4-trimethylammonium-3-(tetradecylcarbamoyl)-aminobutyrate;
R,S-4-trimethylammonium-3-tetradecylaminobutyrate;
R,S-4-trimethylammonium-3-octylaminobutyrate;
R,S-4-trimethylammonium-3-(decansulfonyl)aminobutyrate;
R,S-4-trimethylammonium-3-(nonylsulfamoyl)aminobutyrate;
S-4-trimethylammonium-3-(dodecansulfonyl)aminobutyrate;
R-4-trimethylammonium-3-(dodecansulfonyl)aminobutyrate;

S-4-trimethylammonium-3-(undecylsulfamoyl)aminobutyrate;
R-4-trimethylammonium-3-(undecylsulfamoyl)aminobutyrate;
R-4-trimethylammonium-3-(dodecylcarbamoyl)aminobutyrate;
R-4-trimethylammonium-3-(10-phenoxydecylcarbamoyl)aminobutyrate; and
R-4-trimethylammonium-3-(*trans*- β -styrenesulfonyl)aminobutyrate.

8 35. (New) A process for the preparation of a compound of claim 28,
wherein Z is carbonate (-OCOOR₄), carbamate (-NHCOOR₄), thiocarbamate (-
OCSNHR₄) or thiocarbonate (-OCSOR₄), said process comprising reacting X+
CH₂-CH(OH)-CH₂-Y-, of the desired structure, optionally protected on the acid
Y- group, respectively with an alkyl chloroformate, alkyl isocyanate, alkyl
isothiocyanate or alkyl thiochloroformate, wherein the alkyl moiety is the
desired R₄ alkyl group, to produce the desired compound.

9 36. (New) A process for a preparation of a compound of claim 28,
wherein Z is amide (-NHCOR₄), thioamide (-NHCSR₄), carbamate (-NHCOOR₄),
thiocarbamate (-NHCSOR₄), ureido (-NHCONHR₄), thioureido (-NHCSNHR₄),
sulfinamide (-NHSOR₄), sulfonamide (-NHSO₂R₄), sulfinamoylamino
(-NHSONHR₄), and sulfamide (-NHSO₂NHR₄), said process comprising reacting
X⁺-CH₂-CH(OH)-CH₂-Y-, of the desired structure, optionally protected on the
acid Y- group, respectively with an acyl chloride, thioacyl chloride, alkyl
chloroformate, alkyl thiochloroformate, alkyl isocyanate, alkyl thioisocyanate,
alkyl sulfinyl chlorides, alkyl sulfonyl chlorides, SOCl₂ and alkyl amines, alkyl

sulfamoyl chloride or SOCl_2 and alkyl amine, wherein the alkyl moiety is the desired R_4 alkyl group, to produce the desired compound.

10 37. (New) A process for the preparation of a compound of claim 28, wherein Z is $-\text{OR}_4$ or $-\text{SR}_4$, said process comprising the steps of:

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- (a) reacting a carbonyl compound of formula $\text{Hal-CH}_2\text{-CO-CH}_2\text{-COOR}'$, wherein Hal is a halogen atom and R' is the residue of a suitable ester, with respectively alcohols and thiols R_4OH or R_4SH , to give the respective ketal or thioketal;
 - (b) transforming the respective ketal or thioketal into the respective ether or thioether;
 - (c) substituting the Hal atom with an azido group, and
 - (d) transforming the azido group into the X^+ group to produce the desired compound.

11 38. (New) A process for the preparation of a compound of claim 28, wherein Z is $-\text{NHR}_4$, said process comprising reacting of $\text{X}^+\text{-CH}_2\text{-CH(NH}_2\text{)-CH}_2\text{-Y}^-$ of the desired structure, optionally protected on the acid Y^- group, with alkane carbaldheydes, wherein the alkyl moiety is a one-term lower homologue of the desired R_4 , and subsequent reduction, to produce the desired compound.

12 ³⁹. (New) A pharmaceutical composition comprising a therapeutically effective amount of a compound of claim 28¹, in admixture with a pharmaceutically acceptable vehicle or and excipient.

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cmt 13 ⁴⁰. (New) The pharmaceutical composition according to claim 39¹², wherein an active ingredient suitable for the treatment of diabetes is also present and is selected from the group consisting of sulfonylurea, L-carnitine, fibrate and other agonists of peroxisomal proliferator activated receptor (PPAR- α), HMG-CoA reductase inhibitor, β -sitosterol inhibitor, cholesterol acyltransferase inhibitor, biguanides, cholestyramine, angiotensin II antagonist, melinamide, nicotinic acid, fibrinogen receptor antagonists, aspirin, α -glucosidase inhibitors, insulin secretagogue, insulin and glucagon-like peptides and agonists of PPAR- γ .

14 ⁴¹. (New) A pharmaceutical composition according to claim 39¹², also including an active ingredient suitable for the treatment of obesity selected from the group consisting of fenfluramine, dexfenfluramine, phentiramine, and a β -3-adrenergic receptor agonist.

15 ⁴². (New) A pharmaceutical composition according to claim 39¹², also including an active ingredient suitable for the treatment of high cholesterol levels and in modulating HDL plasma levels, which is selected from the group consisting of fibrates, and other PPAR- α agonists; inhibitors of cholesterol

biosynthesis, HMG-CoA reductase inhibitors, statins, inhibitors of cholesterol absorption, acyl CoA:cholesterol acyltransferase inhibitors, anion exchange resins, nicotinyl alcohol, nicotinic acid or a salt thereof, vitamin E, thyromimetics and L-carnitine.

16 ~~43.~~ (New) A method for treating a subject having hyperactive carnitine palmitoyl-transferase comprising administering to said subject an effective amount of a compound of claim ~~28.~~ 1

17 ~~44.~~ (New) A method for treating a subject having hyperglycaemia, diabetes, heart failure or ischemia comprising administering to said subject an effective amount of a compound of claim ~~28.~~ 1

18 ~~45.~~ (New) A method for treating a subject having obesity comprising administering to said subject an effective amount of a compound of claim ~~28.~~ 1

19 ~~46.~~ (New) A method for treating a subject having high triglyceridemia comprising administering to said subject an effective amount of a compound of claim ~~28.~~ 1

20 ~~47.~~ (New) A method for treating a subject having hypertension comprising administering to said subject an effective amount of a compound of claim ~~28.~~ 1

2) 48. (New) A method of modulating high cholesterol levels or MDL

plasma levels in a subject in need of same, said method comprising

administering to said subject an effective amount of a compound of claim 28.

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